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HEART FAILURE

INTERMEDIATE

CASE REPORT: CLINICAL CASE

A Tale of 2 Aneurysms

Cardiogenic Shock Secondary to Vascular Behçet's Syndrome

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ABSTRACT

A patient with vascular Behçet's syndrome (BS), a subtype of BS with mainly venous/arterial manifestations, presented with a left main aneurysm/thrombus and cardiogenic shock. The clinical diagnosis of BS includes mucocutaneous, vascular, and neurologic criteria. It is important to consider vascular BS as a nonatherosclerotic cause of coronary aneurysms. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1858–1862) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 45-year-old man of Portuguese descent presented with 24 hours of abdominal discomfort and vomiting, followed by severe chest pain radiating to his back. On admission, the patient was in respiratory distress and diaphoretic, although he was awake and moving all four limbs. The mean arterial pressure (MAP) was 68 mm Hg, the heart rate was 104 beats/min, and the oxygen saturation was 88% on 2L nasal prongs. There were decreased breath sounds with diffuse crackles bilaterally. No cardiac murmur was discerned, and there was no peripheral edema. The electrocardiogram (ECG) on presentation to the hospital showed ST-segment elevation in aVR and aVL and ST-segment depression in the inferior and lateral leads (**Figure 1A**). Troponin level on admission was 26,763 ng/L (normal reference value <15 ng/L). His initial symptoms of abdominal pain and vomiting were secondary to hypoperfusion with an initial lactate of 2.2 mmol/L (normal reference value <1.7 mmol/L). Coronary angiography revealed a giant left main (LM) aneurysm measuring approximately 10 mm, with thrombus with Thrombolysis In Myocardial Infarction (TIMI) flow grade 1-2 (**Figure 2,** Video 1). Balloon angioplasty (size 2.5 mm) to the LM temporarily restored flow to the left anterior descending (LAD), intermediate, and circumflex arteries, with resulting TIMI flow grade 2 with a 90% stenosis and aneurysm distal to the obstruction (Videos 2a to 2b). Left ventricular (LV) end-diastolic pressure was 30 mm Hg. Given the worsening instability of the patient's condition, he was intubated, and an intra-aortic balloon pump (IABP) was placed.

LEARNING OBJECTIVES

- To recognize the differential diagnosis for coronary aneurysms identified by coronary angiography.
- To understand the incidence, prevalence, and cardiac manifestations of vascular Behçet's syndrome.

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Because the patient was in cardiogenic shock and there was no feasible percutaneous option to address the thrombotic burden in the setting of an LM aneurysm, the decision was made to take the patient urgently to the operating room instead of delaying surgery. He underwent coronary artery bypass grafting with the left internal mammary artery to the LAD and a saphenous vein graft to the intermediate. An intraoperative transesophageal echocardiogram (TEE) showed a left ventricular ejection fraction (LVEF) of <30% and mild right ventricular dysfunction. His postoperative ECG showed an extensive anterolateral myocardial infarction (**Figure 1B**). Troponin peak was over 50,000 ng/L (normal reference value <15 ng/L). The first postoperative echocardiogram showed an LVEF of 20% to 25%.

Four days postoperatively, the patient remained in refractory cardiogenic shock, requiring high doses of milrinone, nitric oxide, and vasopressors. His cardiac index was 2.1 mL/min/m^2, right atrial pressure was 23 mm Hg, pulmonary capillary wedge pressure was 30 mm Hg, and systematic vascular resistance was dynes·s/cm^5. Inasmuch as he remained mechanically ventilated and the IABP was still in place, and he required 3 vasopressor agents and milrinone, he was transferred to an advanced heart failure center for consideration of mechanical circulatory support (MCS).

MEDICAL HISTORY

His medical history included a history of unprovoked deep venous thrombosis (DVT) and pulmonary embolism (PE) at age 30 years without evidence of hypercoagulability. He was not taking any medications. He was a former smoker and occasionally used cannabis.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of a coronary aneurysm includes atherosclerosis, congenital malformations, connective tissue disease, and inflammatory disorders, including sequelae of childhood-onset Kawasaki disease, and vasculitis, such as polyarteritis nodosa and vascular Behçet's syndrome (BS).

INVESTIGATIONS

Antinuclear antibody, rheumatoid factor, anticyclic citrullinated peptide, and antineutrophil cytoplasmic antibodies were all negative. Anticardiolipin antibody was positive; however, B2 glycoprotein was negative. The patient was found to be a carrier of the human leukocyte antigen (HLA)-B51 allele. There was no history of childhood illnesses suggestive of Kawasaki disease, oral or genital ulcers, or mucocutaneous manifestations on physical examination.

Despite receiving therapeutic anticoagulant agents, he received a diagnosis of bilateral segmental and subsegmental pulmonary emboli, and occlusion of the common hepatic artery proper with porta hepatis arterial aneurysm measuring 6 mm (**Figures 3A to 3B**).

MANAGEMENT (MEDICAL/INTERVENTIONS)

He underwent cannulation onto peripheral venoarterial extracorporeal membrane oxygenation (VA-ECMO) as a bridge to decision. Intraoperative TEE showed severe biventricular dysfunction, smoke in the LV, and early left atrial appendage thrombus (Supplemental Figure S1). Balloon septostomy for LV venting was performed. Subsequent echocardiograms reported LVEF measurements of <10%.

In this male patient of Portuguese descent, who was a carrier of the HLAB51 allele, with coronary and hepatic aneurysms, and venous thromboembolism, vascular BS was considered the most unifying diagnosis. Intravenous solumedrol was initiated. Cyclophosphamide was considered but was not given because of *Escherichia coli* bacteremia.

DISCUSSION

We describe a case of a young man who received a diagnosis of vascular BS after presenting with a giant LM coronary artery aneurysm with thrombus and cardiogenic shock requiring VA-ECMO support. BS is classified as a chronic relapsing inflammatory disorder with often widespread involvement in multiple organ systems (1). The cause of BS is not entirely known, although infectious, environmental, genetic, and immunologic factors may be contributory (2). Vascular BS is a subtype of BS with predominantly vascular manifestations. There is no pathognomonic test for the diagnosis of vascular BS, and any size and type of vessel can be involved (1,3).

The prevalence of vascular BS varies between series. Vascular manifestations have been reported in 2.2% to 50% of the affected populations, and a predominance of venous over arterial involvement, with 1 study reporting 84.6% of vascular BS patients having only venous disease, 8.1% only arterial, and 4.2% having both (1,4). Venous involvement includes superficial phlebitis, DVT, the superior and inferior vena cava, and Budd-Chiari syndrome (1,4,5). The

ABBREVIATIONS AND ACRONYMS

HLA = human leukocyte antigen

IABP = intra-aortic balloon pump

LAD = left anterior descending

LM = left main

MAP = mean arterial pressure

MCS = mechanical circulatory support

TEE = transesophageal echocardiogram

TIMI = thrombolysis in myocardial infarction

VA-ECMO = venoarterial extracorporeal membrane oxygenation

vascular BS = vascular Behçet's syndrome

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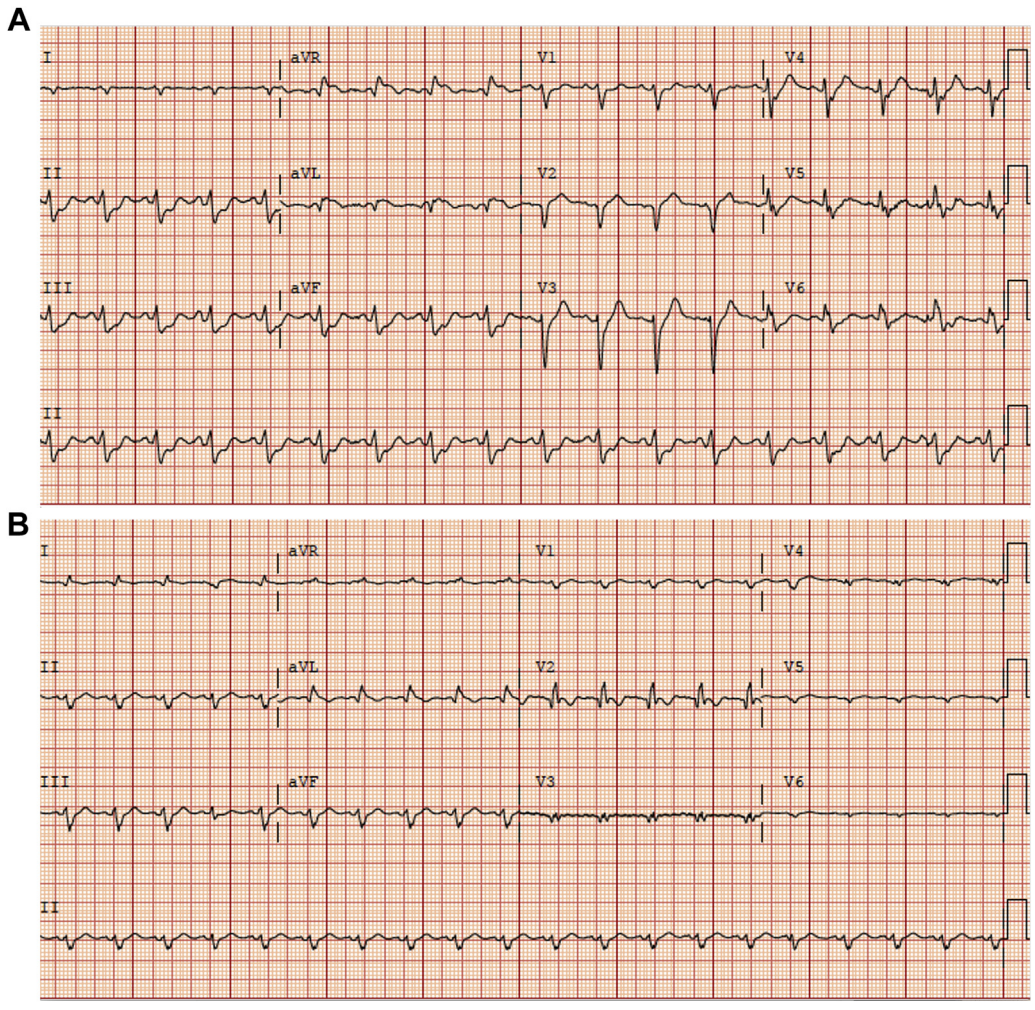
FIGURE 1 Initial and Postoperative Electrocardiograms



(A) Electrocardiogram on presentation to hospital. There is ST-segment elevation in aVL and aVR and ST-segment depression in the inferior and lateral leads. **(B)** Postoperative electrocardiogram. There is an extensive anterolateral myocardial infarction.

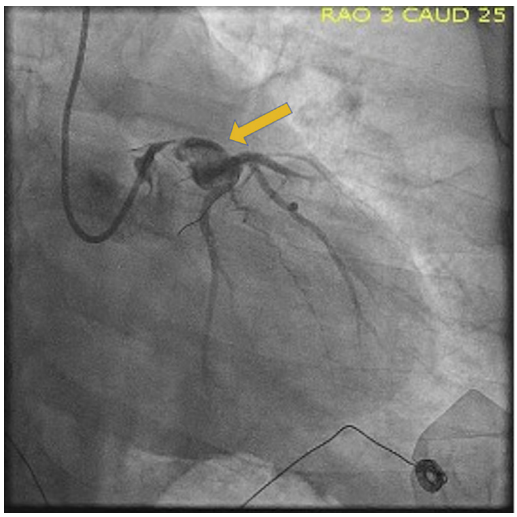
frequency of arterial involvement varies between studies, ranging from 0.7% to 14% depending on the diagnostic methods and level of investigations performed (5,6). Arterial manifestations include aneurysm formation (47.3%), occlusion (36.5%), stenosis (13.5%), and aortitis (2.7%) (7). The most common vessels involved are the aorta, femoral, pulmonary, and iliac arteries. Coronary artery involvement is rare. In a study of 52 patients with BS with cardiac manifestations, 1.9% had a documented coronary aneurysm (8). Arterial involvement in BS has been shown to be an independent risk factor for death (6).

Further research is needed regarding the management of vascular BS. Immunosuppressive therapy is the cornerstone of treatment because the primary pathologic feature is inflammation of blood vessels (9). Corticosteroids, azathioprine, cyclophosphamide, and cyclosporine A are suggested for venous thromboembolism. Cyclophosphamide and corticosteroids are recommended for the treatment of peripheral and pulmonary artery aneurysms (9). Advanced heart failure therapies have been described in cases with aortic involvement (10). To our knowledge, this is the first report of MCS for vascular BS with coronary artery involvement.

FOLLOW-UP

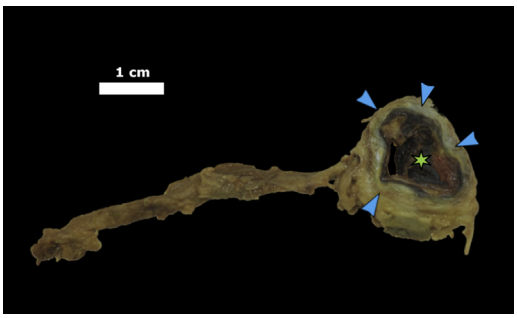
Although there was sufficient biventricular recovery to decannulate from VA-ECMO, the patient died of

FIGURE 2 Coronary Angiography on Admission



The angiogram shows the left main coronary artery aneurysm **(yellow arrow)** and thrombus with no significant atherosclerosis otherwise.

FIGURE 4 Postmortem Pathological Findings



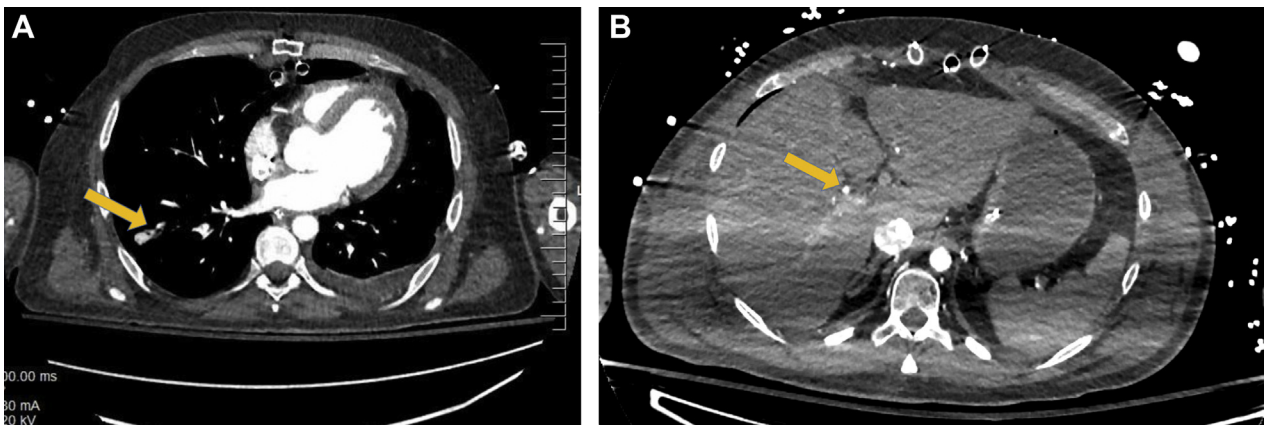
Autopsy specimen of the left main coronary artery aneurysm. To the right is markedly thinned, fibrotic, and focally calcified vessel wall **(blue arrowheads)** with extensive luminal thrombus **(green star)** (**dark red**, unorganized thrombus; **off-white**, organizing thrombus). The left anterior descending artery is seen extending leftwards from the aneurysm. Scale bar for size included.

sepsis. The postmortem findings included a 3-cm LM coronary artery aneurysm (Figure 4) with acute myocardial infarction and adventitial inflammation along with segmental PE. The findings supported a recent vasculitis, and the leading diagnosis remained vascular BS.

CONCLUSIONS

Vascular BS is a clinical diagnosis and in rare circumstances may initially present with coronary artery aneurysms. This case highlights the importance of early clinical suspicion of vascular BS in young patients with coronary artery aneurysms and the need

FIGURE 3 Computed Tomography Scans During Admission



(A) Computed tomography of the chest. Bilateral pulmonary emboli are seen **(yellow arrow)**. **(B)** Computed tomography of the abdomen. Porta hepatis arterial aneurysm measuring 6 mm **(yellow arrow)**.

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to evaluate for both venous and other arterial involvement.

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KEY WORDS acute coronary syndrome, cardiac assist devices, systolic heart failure

APPENDIX For a supplemental figure and videos, please see the online version of this paper.